

CLINICAL VIGNETTE

Classic Kaposi's Sarcoma Presenting with Edema: A Case Report

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Case Report

A 67-year-old Russian immigrant has been followed for hypertension, hyperlipidemia and venous insufficiency for 7 years. After a left hip fracture, the patient developed increasing lower extremity edema, left greater than right. He had been using a beta blocker, clonidine, amlodipine and an ace inhibitor to control his hypertension. He was also taking warfarin in the post operative period for DVT prophylaxis. The swelling was thought to be secondary to hypertension medication and venous insufficiency exacerbated by prolonged immobility following surgery. A diuretic was added with little effect. Bilateral lower extremity duplex scans were performed, which showed no sign of deep vein thrombosis. Venous insufficiency scans revealed minimal regurgitation.

During this period, the patient had noted a non-pruritic rash of the lower extremities. Starting at the lateral aspect of the lower calf to the mid thigh, he developed a dark brown discoloration in patches of 2-3 cm in length. He complained of tenderness at these areas, particularly with his prescribed compression stockings. The initial lesions were thought to be related to venous stasis skin changes. However, with expansion of the lesions, a dermatology consult was obtained, and he underwent biopsy of one of the skin lesions. The biopsy revealed Classic Kaposi's Sarcoma.

Over the ensuing 2 months, he developed an increasing number of lesions over his legs and buttocks with increasing discomfort and edema. He was evaluated by oncology and began systemic chemotherapy with subsequent improvement in his lesions.

Discussion

Classic Kaposi's Sarcoma is a rare angioproliferative disease affecting mainly middle aged to elderly men of Mediterranean or Eastern European descent. The skin lesions of Classic Kaposi's Sarcoma are typically

violet, red or brown and affect the lower extremities. The size of the lesions are quite variable, ranging from a few millimeters to several centimeters¹. As was the case in this patient, concurrent edema is usual, but is due to lymphatic obstruction from the infiltrating skin lesions, rather than venous insufficiency.

Diagnosis is based on identification of the characteristic skin lesions and subsequent biopsy. On biopsy, the histology shows abnormal lymphatics and immunostaining identifies Human Herpes Virus 8, which is a requisite for the diagnosis².

The clinical course of Classic Kaposi's Sarcoma is typically a gradual progression of skin lesions and edema. Mucous membranes can also be involved--typically those of the stomach, esophagus and small intestine. In contrast to AIDS related Kaposi's Sarcoma, though, GI manifestations are less common, usually less than 10% of cases³. Nonetheless, anemia in a patient with Classic Kaposi's Sarcoma warrants endoscopic evaluation. While the majority of cases do not result in significant morbidity or mortality, a small subset can present with a rapidly progressive course (in one case study, only 2% of patients followed died of disseminated Kaposi's Sarcoma)⁴.

Treatment for Classic Kaposi's Sarcoma varies, largely due to the rarity of the disease. Local disease with limited functional loss can simply be observed. Often treatment is aimed at symptom control--cryotherapy or dermatologic surgery for irritated lesions and compression hose to manage lymphedema. For more advanced disease, treatments include radiation therapy, and intralesional therapy with chemotherapeutic agents⁵. Topical agents include Imiquimod and retinoic acids which have proven effective for treatment of symptomatic lesions. In patients who have rapidly progressive disease, visceral organ involvement or severe functional decline, systemic chemotherapy is

indicated. Response rates to systemic therapy are typically very good with one study showing a 71% response rate to doxyrubicin. Anti-angiogenesis and immunomodulatory agents, such as Thalidomide and interferon alpha have also been used in treating advanced disease or in patients who have failed chemotherapeutic treatment⁶.

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